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Multiple Subperiosteal Sarcoma of the Skull,

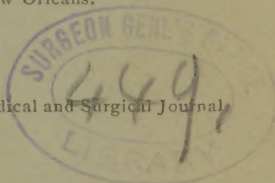
ASSOCIATED WITH

Necrosis of Vertical Plate of Frontal. Extirpation of
Necrotic Vertical Plate and Tumors—Recovery.

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Visiting Surgeon, Charity Hospital, New Orleans.

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presented by the author.

Multiple Subperiosteal Sarcoma of the Skull,

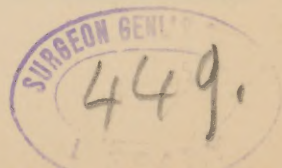
Associated with Necrosis of Vertical Plate of Frontal. Extirpation of Necrotic Vertical Plate and Tumors. Recovery.

By **RUDOLPH MATAS, M. D.**, Visiting Surgeon, Charity Hospital, New Orleans.

The peculiar features presented by this case—viz.: the unusual extent of the necrosis, involving the whole vertical portion of the frontal; its association with sarcomatous formations in other parts of the skull; the doubtful cause of the necrotic process, and, finally, the operation required for the removal of the diseased areas—justify, I believe, a report of this case and its presentation to you.

Observation.—Ephraim B. Hicks, colored, laborer, æt. 35 years, was admitted in Ward 2, Charity Hospital, March 3, 1888. His parents are both dead and he has no recollection of any hereditary disease in his family, especially no history pointing to tuberculosis, syphilis or cancer. He himself denied having acquired the syphilitic taint, and on questioning gave no evidence that positively indicated the existence of the disease prior to the present trouble.

History.—He stated that on June 1, 1882, seven years ago, he noticed a small “knot” on the right side of the forehead, close to the temple, at the site of a scar, which had resulted from a recent blow. This remained indolent until last year in July, eight months before admission, when the tumor began to swell rapidly. It became painful, was poulticed for a long time, because it was so hard, and was finally opened by some medical man who saw him. After the cutting, the lump “festered,” ulcerated and finally “eat away” the soft parts of the forehead, leaving him in the pitiable condition which he presented on the day of admission. During his illness, prior to this time, he had been seen by several physicians in the country, who

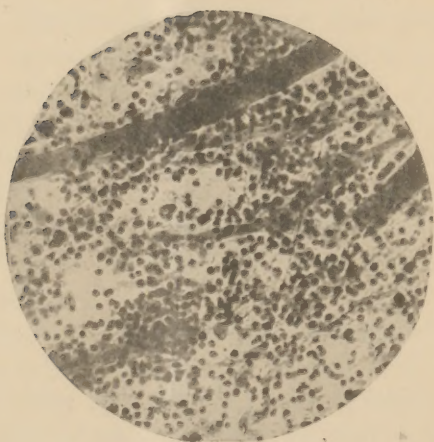


regarded his disease as a syphilitic manifestation and treated him accordingly. About a week before seeking admission into the Charity Hospital, he noticed two other swellings similar to the first, both about the size of pigeons' eggs and appearing over the parietal regions.

Condition on Admission.—The right side of the forehead presented a large oval surface, measuring over three inches and a half in longest diameter and slightly less in the opposite, over which all the soft parts had been completely ulcerated away. The edges of this area were hard, inverted and of a dark bluish cicatricial appearance. They sharply defined externally an area of *denuded* bone, plainly necrosed, which was rough, riddled with small foramina and irregular crevices, from which an exceedingly foul and offensive greyish pus continually oozed. The discharge of pus was most plentiful in the lower or most dependent portion of the ulcerated surface, the pus here trickling continually downwards by large drops upon the face. A probe was passed readily in all directions under the thickened integument which formed the edges of the ulcer, and detected rough and denuded bone all over the area of the frontal, the necrosis being limited laterally by the frontal portion of the temporal ridge on the left side and encroaching considerably into the temporal fossa on the right.*

At the time the patient presented himself for advice in my service the right parietal tumor was longer than the left; it felt slightly larger than a pigeon's egg, was soft, and on close examination gave a deceptive feeling of semi-fluctuation. It was evidently fixed and immovable, and gave the impression of being fixed to the periosteum or skull. This tumor was situated on the vertex, at a point one and a half inch from the bregma and two inches to the right of the median line.

*Figure I, copied from a photograph, illustrates the exceeding abundance of the secretion by the whitish streak which is seen running from the ulcer to the right eye-brow, and which could not be kept dry even for ordinary photographing. The drainage tube inserted under the upper margin of the ulcer indicates its undermined character, and the prominence of the head over the right parietal eminence indicates the site of the right parietal sarcoma.



Another tumor, smaller than the above and like it completely invested by the scalp tissues, was situated at a point two and a half inches back of the bregma and two inches to the left of the median line.

As to the general condition of the patient we may say that he was evidently very anæmic; that he was emaciated, and rapidly becoming more so, and would soon sink into complete marasmus if not relieved, as the drain on his resources from suppuration, pain, lack of rest and unhappiness from the increasing misery and repulsiveness of his condition was constantly telling upon him. His pulse was full, easily excited, but not febrile; his temperature normal, but he complained of constant and considerable pain under the forehead, where the disease was situated, and lately he staggered occasionally with a dizziness amounting almost to vertigo.

These last symptoms gave us much apprehension as to the future possibility of meningeal involvement and secondary brain troubles, and they especially urged interference, though it was possible that they might be due to the general anæmic condition.

Diagnosis.—A most important consideration that now presented itself before proceeding further with the management of the case was the diagnosis. What was the cause of the necrosis? and what was the nature of the two parietal growths which the patient stated were in every way analogous to the primary and much larger necrotic disease of the forehead?

The possible causes that immediately suggested themselves were: 1, syphilitic osteo-periostitis, with tertiary gummata of the cranium; 2, tubercular osteo-periostitis of frontal, with tubercular deposits over the parietals; 3, primary neoplastic (sarcomatous) deposits of cranial vault.

Notwithstanding the absolutely negative history and the failure of the anti-syphilitic treatment applied by other practitioners, I decided to give the patient the benefit of any

doubt in this direction, and to notice the effect on the growing parietal tumors of a positive anti-syphilitic medication. He was therefore put at once on the following: R̄ Hydrarg. biniodidi, gr. i; potass. iodidi, ʒss; syr. ferri iodidi, ʒiii; syr. zingiber., q. s. ad ʒviii M. et S.: Tablespoonful four times daily. At the same time a tonic pill of strychn. sulph., acid arsenios., quin. sulph., was administered *ter in die*. Local sublimate dressings 1:2000, with carbolic acid 2½ per cent., were kept constantly applied over the denuded and necrotic surface.

The patient took the anti-syphilitic mixture well, but his headache and dizziness gradually increased, and the ulcerated forehead was as purulent and offensive as ever. This was a case which appeared to defy all antisepsis. Even iodoform applied plentifully, in addition to frequent bichloride and carbolic irrigation and thorough drainage failed to neutralize the fetor. This difficulty, in controlling the discharge and bad odor, was due to the fact that owing to the extensive undermining of the scalp the antiseptic dressings could not be made to act effectually.

Shortly after placing the patient on the preceding specific treatment I explored the doubtful semi-solid tumors of the parietal regions with an exploring syringe. Nothing but a little blood was drawn through the needle, which was large and pierced the whole thickness of the mass. This exploration appeared to indicate the solid character of the swellings, and reduced the diagnosis to either (1) a syphilitic gumma, or (2) neoplastic deposit. In the meantime the anti-syphilitic treatment was unsparingly continued, and as there were no signs of iodism this appeared also *to confirm the syphilitic diagnosis*.*

*The tolerance of the potassium iodide in this case has some bearing on the discussion which has recently arisen between Drs. H. C. Wood and J. William White on the value of the iodides in establishing the differential diagnosis of syphilis in obscure cases. Dr. H. C. Wood stated some time ago that "in all cases of doubtful diagnosis of cerebral syphilis the so-called therapeutic test should be employed, and if 60 grains of the iodide of potassium per day fail to produce iodism, for all practical purposes the person may be considered to be syphilitic."

The subsequent histological examination of the tumor and history of the case proved that this patient was not syphilitic, and still he tolerated the iodides admirably, since he took for over 4 weeks exactly 60 grains a day. Another patient, a tailor, suffering with a well-marked and most distressing aneurism of the ascending arch of the aorta has taken 40 grains three times daily during the last two years; he is not a syphilitic subject, but

On the other hand, the fact that no improvement, no favorable impression was being made upon the patient's condition, either locally or generally, but, on the contrary, that the patient was progressing unfavorably, led me to test the secretions of the ulcer for bacilli tuberculosis. The pus and débris from the forehead were very carefully and repeatedly examined, but outside of the existence of the usual cocci and microörganisms of suppuration, no *bacilli tuberculosis* were found,

The absence of tubercular bacilli did not, of course, exclude the diagnosis of tuberculosis, since almost all observers have proved that the typical bacilli may be absent from pus and still exist in demonstrable quantities in the granulation tissue (König, Volkman, Schuchardt and Krausse, Roswell Park and others.) Only the inoculation test would have been decisive, but this was impracticable at the time, and the diagnosis consequently remained doubtful, both as to the syphilitic or tubercular origin of the disease.*

was once a very hard drinker, and he has never shown a sign of iodism, no coryza, epiphora or even the acnoid eruption. To this tolerance for the iodides I would ascribe much of the decided benefit to, and almost cure of, his aneurism. These two cases would, therefore, tend to support Dr. White's position, who dissents from Dr. Wood's views, and has recently secured the opinions of a number of eminent syphilographers and general practitioners, who in their letters coincide with Dr. White and express themselves decidedly against the position taken by Dr. Wood. [See *Therapeutic Gazette* of March, 1889, and editorial in *N. Y. Medical Record* of May 4, 1889.] While some admit that in tertiary syphilis patients as a rule stand large doses of iodide very well, the exceptions to this rule are too numerous to make it of any diagnostic value. The general opinion seems to be that tolerance of iodides is a matter of personal idiosyncrasy, and is not modified by syphilis.

"In commenting upon the letters Dr. Wood modifies somewhat the statement we have first quoted, and says that in advanced syphilis iodide of potassium is usually tolerated much more freely than in health, and that this rule or coincidence is of sufficiently wide application to be of value in prognostication and treatment."

Since the first appearance of Dr. Wood's opinion I have always believed it true, and notwithstanding contradictory cases that I have noted, I am still convinced that there is much that is true in it; though I would lay less stress upon the diagnostic importance of the "therapeutic test" than formerly. It is a question which is to be decided not by mere opinion only, but recorded observation.

*The last possibility—i. e., that the two parietal tumors were neoplastic deposits and the necrosis of the frontal also of malignant origin—I could not decide except by histological examination of the tissues, and this could not be done until after extirpation.

In connection with the inoculation test for tubercular lesions, it is instructive to note the work of Cavel of Berne, which is referred to by Prof. N. Senn in both of his recent and admirable books, viz.: "Four Months Among the Surgeons of Europe," 1887, Chicago, pp. 154; and his later, "Surgical Bacteriology," Philadelphia, Lea Bros. & Co., 1889, pp. 191. Cavel has been studying in a systematic manner the diagnostic value of implantation of tubercular material in animals, mainly Guinea pigs. Granulation tissue from tubercular joints in his experiments on Guinea pigs invariably produces acute, diffuse tuberculosis, and death in from five to six weeks. The course of the disease in the animal is typical. At the point of inoculation a hard nodule appears first, the result of a traumatic inflammation of the tissues around the graft. Next a lymphatic gland becomes enlarged in the immediate vicinity of the primary seat of infection, which is always done in the flank, consequently the inguinal glands enlarge first; glandular infection increases rapidly. After the whole chain of lymphatic glands in the groin are involved, the axillary glands become affected.

Again, though the history appeared distinctly favorable to the theory of sarcomatous diseases, still it did seem probable, that if the bone disease (frontal) had been of cancerous or sarcomatous origin it should have given rise to an osteophytic growth, which would have infiltrated and incorporated itself in the cranial vault instead of limiting its invasion to the periosteum, and leaving a large and well-defined denuded surface. Anyway, the satisfaction of a positive or definite diagnosis, outside of the simple fact of the necrosis, was reluctantly abandoned, and it was decided to make an exploratory operation, which would expose the diseased area as completely as possible and then if practicable allow the removal of the whole by a radical operation.

Accordingly, on April 4, 1888, after shaving the scalp and subjecting the parts to a thorough antiseptic preparation, the patient was placed under chloroform. A transverse incision towards the left temple, as shown in figure 2, was carried to the left temporal fossa through all the scalp tissues, to the bone. This incision, after a little dissection allowed the writer to recognize the fact that the necrosis extended to about half an inch of the coronal suture above, and that a well-marked line of demarcation existed. Encouraged by this fact, the whole skin of the forehead was completely and readily detached, owing to the thickness of the periosteum, due to chronic periostitis. The whole soft forehead was then reflected downwards over the eyes; a vertical incision was then added, which allowed a lateral flap to be detached, and permitted the whole necrotic area to be completely and thoroughly exposed. It was then seen that the highest point reached by the dead bone was

At the post mortem examination it was always found that of the internal organs the spleen becomes affected first, then the liver and lungs, but usually the disease is so diffuse that scarcely an organ remains exempt. When the diagnosis between syphilis and tuberculosis cannot be made, either clinically or by use of the microscope, inoculation experiments always give positive and reliable information. When it is syphilitic the inoculation is harmless, and the animal remains well. If tubercular, it dies in the time noted, five or six weeks at the furthest, and very exceptionally five months. The tubercle bacillus is invariably found in the contents of abscesses or in the diseased tissues. Kocher of Berne, in whose laboratory these experiments have been conducted, has learned to depend upon the test as the only decisive one in doubtful cases.

about one inch from the bregma, and the lowest about one-quarter inch from the superciliary ridges; laterally it reached the temporal fossæ.

An attempt was now made to elevate the large and well defined sequestrum, by introducing an elevator between the healthy right parietal and the dead bone, but it proved ineffectual. The crown of a large Galt's trephine was applied over the line of demarcation, so that the disc that was to be removed would include both a segment of the sequestrum and of the healthy bone. The trephine was carried to the vitreous plate, almost to the dura mater; and then after the removal of the trephined disc another attempt was made to elevate the sequestrum by using the healthy bone as a fulcrum; but again the attempt failed, because of the strength of the attachment between the sequestrum and its bed. Several (three) approximated discs of bone were removed at different spots along the line of demarcation not with the Galt, but with a larger cylindrical trephine. But the attempts at elevation still failed; and seeing this I decided to loosen the peripheral attachments of the sequestrum by means of the chisel. With the help of this instrument the line of demarcation was enlarged at its most distinct portion above, and the chisel (a large carpenter's instrument with a long handle) was finally insinuated below the dead bone, and with the help of the healthy bone as a fulcrum, it was not long before the sequestrum yielded with a very distinct report, indicating the firmness of the connections of the necrosed fragment. As the sequestrum was detached it was observed that the area of skull occupied by it was filled with a large crop of granulations, which were bathed in some places with pus, and were studded here and there with new osteophytic islands. It was furthermore discovered that the sequestrum involved the whole thickness of the skull only in its middle portion in the centre of the forehead, over the longitudinal sinus, where the dura mater visibly pulsed. In the peripheral parts the necrotic plate extended

only to the vitreous. It involved, throughout, the external table and the diploë, and in size represented the major portion of the whole vertical plate.

Volkman's curette was than liberally applied over the whole granular *sub-necrotic* territory, taking care not to scrape too vigorously over the soft pulsating dura mater, which was largely exposed in the median line. The flaps of the scalp and forehead were then pared at the edges, which were much thickened, and replaced in their original positions.

An incision was now carried from the forehead to the right parietal tumor. The tumor was thereby exposed and found to be a solid tumor of a slightly fibromatous consistence: it seemed to be softened in spots. This tumor was removed with the periosteum to which it was attached, and the bone under it scraped with a chisel.

The left parietal was likewise thoroughly removed by a separate incision.

The wound was irrigated during the operation with a stream of acid bichloride solution (Laplace's) 1:2000, and after its termination, when the flaps were readjusted a dressing of alternate layers of bichloride and iodoform gauze were applied to the head.

The patient was greatly prostrated by the operation, which lasted over one hour, but he rallied promptly, so that in four days he was able to sit up out of bed. The removal of the large sequestrum and the almost complete arrest of all the wasting suppurative drain immediately told, and most beneficially, on the patient's condition. His weight increased; he slept perfectly; all the head symptoms disappeared, and by April 14, 1889, ten days after the operation, the patient was discharged by his own request. The wound had not yet healed completely, but was practically on the eve of complete cicatrization, excepting over the large exposed oval, uncovered by skin, which had to heal by granulation.

It was noticed that as the process of healing advanced,

islands of calcareous material were formed in little spots over the whole field, and even the places where the dura had been felt distinctly pulsating gradually became hard and finally ceased to pulsate altogether.

As the granulation cicatrized the new skin (cicatricial) became studded with these osseous nodules, which were quite prominent and could be easily felt. In fact, when the patient returned, three months after the operation, when we had his photograph taken (fig. 2) these osseous islands could be seen as well as felt, and they evidently assisted very materially in protecting the otherwise purely membranous covering which the brain had at this point.

Returning now to the nature of the trouble, we will state that both the parietal tumors and slices of the edge of the frontal ulcer were submitted to the pathologist of the hospital, Dr. Schmidt, who, after a careful examination, pronounced all the specimens sarcomatous, round and spindle-celled. Some time afterwards a section of one of the parietal tumors was sent to Dr. Gray, microscopist of the medical museum at Washington, who photographed the slide, which is here appended. This negative illustrates a portion of the tumor which had undergone myxomatous degeneration. The section was a little thick, but it was admirably managed by Dr. Gray, who has certainly reproduced this portion of the field with great fidelity.

The diagnosis in this case was therefore neither syphilis nor tuberculosis of the cranium, but sarcoma. I must confess that though this diagnosis cannot be questioned in regard to the two small tumors removed over the parietals, it is still a matter of grave doubt in my mind if the extensive necrosis of the frontal was the result of neoplastic ulceration, or was the consequence of an osteo-periostitis of simple traumatic origin, or one resulting from specific infection, such as tubercle, etc. The limitation of the neoplastic infiltration, if such existed, to the periosteum, and the non-incorporation of the bone by the neoplasm, is something

truly exceptional and cannot be explained unless it be in the light of a very rapid desintegration of the soft supra-periosteal parts—a sloughing of these parts before the neoplasm had an opportunity to infiltrate the underlying bone.

Anyway, whether this case exemplifies a mixed type of cranial disease or a pure one, it teaches the difficulties attending the establishment of a clear and definite diagnosis in some cases, and the propriety of giving the patients the benefit of the doubt, and treating them, as if they were malignant by thorough exploration and eradication.